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Letter to the Editor

Transient Neonatal Arthrogryposis: Another Case

To the Editor:

In 1986, one of us [Robinow, 1986] described monozygotic diamniotic twin girls with the twin-twin transfusion syndrome. At birth, twin A, the transfusion recipient, had polyhydramnios of 2,000–3,000 ml. Her weight was 2.58 kg. She was in cardiac failure (tachypnea, hepatomegaly, gallop rhythm and generalized edema). An echocardiogram showed increased thickness of the septum and the left ventricular wall. Her muscles were unduly firm and rigid. Knees, hips and elbows could not be fully extended. WBC were 55,000, SGOT was 355 units, SGPT 1,005 units, CPK 2,200 units. Within 3 hours after birth, she voided more than 180 ml and after a few more hours all signs of cardiac failure had resolved. A week later the muscles had become softer and the flexion contractures had almost completely disappeared. SGOT had decreased to 36, SGPT to 71, and CPK to 21 units. She seemed to have fully recovered, but a few weeks later she was found to have mild spastic diplegia. Twin B, the transfusion donor, weighed 1.93 kg at birth and was asymptomatic.

CLINICAL REPORT

This infant boy was delivered at term by C-section because his mother had large uterine fibroids. There was neither oligo- nor polyhydramnios. Fetal movements were reported as having been active but the cord was unduly short. Birth weight was 2.63 kg, length 51 cm, OFC 31.5 cm. Apgar scores were 5 and 8 after 1 and 5 minutes, respectively. Abnormal findings were a 2-vessel cord, nuchal edema, one undescended testicle, a mild, unclassified skeletal dysplasia, bilateral club feet and flexion contractures of hips, knees and elbows. The muscles were unduly firm. The fingers were tightly overlapping, somewhat resembling those in the 18 trisomy syndrome. The karyotype was 46,XY. On the day after birth, CPK was 1,520, SGOT 56. Six days later, the CPK had decreased to 172, the SGOT to 21. Cranial ultrasonography gave normal results. At age 5 weeks, the contractures had become distinctly less severe and the club feet had substantially improved without casting.

DISCUSSION

These two infants share perinatal disturbances of muscle with transient elevations of muscle enzymes, in a distribution suggestive of amyoplasia. The pathogenetic mechanism is unknown. The short umbilical cord in case 2 suggests the fetal akinesia syndrome. Thus, different mechanisms appear to produce the picture of neonatal arthrogryposis.

REFERENCE

Robinow M (1986): Transient neonatal arthrogryposis: A presumed sequel of antenatal hypoxia. Am J Med Genet 25:167–168.

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